Br Heart J 1982; **47**: 35–40

Atresia of left atrioventricular connection
Surgical considerations

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SUMMARY We have studied 17 patients diagnosed in life as having absent left atrioventricular connection (14 patients) or an imperforate left atrioventricular valve (three patients). Ten patients presented below the age of 3 months with tachypnoea, mild cyanosis, and heart failure. Seven patients, including two with moderate and one with severe pulmonary outflow tract obstruction, presented at a mean age of 45 months (range six months to 16 years). Six patients had a non-restrictive interatrial communication when first catheterised. In the remainder the mean interatrial gradient was 14 mmHg. Excluding patients with pulmonary outflow tract obstruction the mean pulmonary vascular resistance was 6·7 units/m^2. Patients were managed by balloon atrial septostomy (four patients), atrial septectomy (nine patients), pulmonary artery banding (nine patients), systemic pulmonary anastomoses (one patient), and a modified Fontan's procedure (one patient). There were five hospital deaths. Two followed pulmonary artery banding and three followed atrial septectomy. The mean length of follow-up in the surviving patients is $5\frac{1}{2}$ years (range five months to 15 years). The six patients who had an atrial septectomy, and one patient who had a modified Fontan's procedure, are well palliated with minimal reduction in exercise tolerance and mild cyanosis at rest. Three patients, one with an atrial septostomy, one who had pulmonary artery banding, and one who had no surgical procedure have evidence of pulmonary vascular disease. A further patient who had a balloon septostomy has electrocardiographic evidence of left atrial hypertrophy. We conclude that atrial septectomy plus or minus pulmonary artery banding provides good palliation and prevents the development of pulmonary vascular disease. A modified Fontan's procedure should be considered as an alternative or subsequent procedure in patients with suitable anatomy.

Atresia of the left atrioventricular orifice in atrial situs solitus is a rare congenital heart defect.[1–3] The majority of cases also have aortic atresia with a correspondingly poor prognosis. Those cases which have a patent aortic root have a better prognosis,[4 5] the longest survival without palliation being in those with an associated non-restrictive interatrial communication and mild to moderate pulmonary stenosis.[6] In this report we describe our experience with the surgical management of patients with absent left atrioventricular connection or imperforate left atrioventricular valve in situs solitus, a topic recently discussed elsewhere.[7] We used palliative procedures designed to relieve left atrial hypertension and to render pulmonary blood flow optimal. In addition we were able to perform a successful modified Fontan's procedure in one case.

Accepted for publication 19 June 1981

Patients

Fourteen patients with absence of the left atrioventricular connection and three patients with an imperforate left atrioventricular valve with patent subaortic outflow tract were seen at the Brompton Hospital between January 1964 and June 1980. Absent left connection is diagnosed when there is no communication, either actual or patential, between the floor of the left atrium and the underlying ventricular mass. The atrial floor is completely separated from the ventricle by atrioventricular sulcus tissue (Fig. 1a). We separated these hearts from those in which an imperforate membrane was interposed between the left atrium and the ventricular mass, these two structures therefore being potentially connected (Fig. 1b). The presence or absence of a valve membrane with accompanying hypoplastic subvalvar

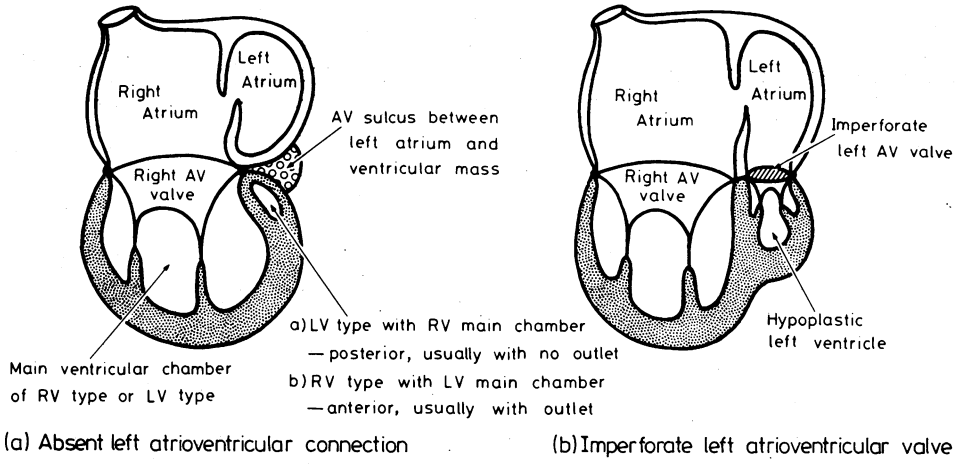

Fig. 1 *(a) Absent left atrioventricular connection. Note atrial floor is completely separate from the ventricle by atrioventricular sulcus tissue. (b) Imperforate left atrioventricular valve in which the left atrial and ventricular cavities are potentially connected.*

apparatus has been subsequently confirmed in all surviving patients by echocardiography.

CLINICAL PRESENTATION

Ten of the 17 patients presented in the first three months of life. Nine of these had tachypnoea, mild cynosis, and congestive cardiac failure, and the tenth had moderate cyanosis. Seven patients presented between the age of 6 months and 16 years. Symptoms in this later group consisted of either increasing cyanosis, decreasing exercise tolerance, or failure to thrive.

CARDIAC CATHETERISATION

Five patients had serial cardiac catheterisations and the total number of cardiac catheterisations performed was 26. Data from these are presented in Table 1. Six patients had an interatrial gradient of less than 5 mmHg at the time of their first catheterisation. Excluding those with a non-restrictive atrial septal defect, two of the patients who had an interatrial gradient of less than 5 mmHg at the time of their first catheterisation have been recatheterised, three and seven years later. In one the interatrial gradient had risen to 13 mmHg and in the other it was not possible to enter the left atrium. Two patients had evidence of moderate and one patient of severe pulmonary outflow tract obstruction. The mean pulmonary vascular resistance, excluding those patients with pulmonary outflow tract obstruction, was 6·7 units/m^2. One patient had evidence of subaortic obstruction.

ANGIOGRAPHY

Selective left atrial injections were performed in all but two patients and confirmed that there was no direct communication between the left atrium and the ventricular mass. The left atrium was visualised on the follow-through in one of the remaining patients and in the other the diagnosis was confirmed at operation.

In the cases with absent left atrioventricular connection, the right atrium was connected via the right atrioventricular valve to a left ventricular chamber in seven patients, a right ventricular chamber in four patients, and a single chamber of indeterminate morphology in three patients. All patients with main chambers of left ventricular type also possessed anterior rudimentary chambers of right ventricular type. In one of these the rudimentary chamber was right sided and supported the pulmonary trunk. In the remaining six the right ventricular chamber was either directly anterior or left sided and supported the aorta.

Rudimentary chambers of left ventricular morphology were present in all four patients with main ventricular chambers of right ventricular type. In all the left ventricular chamber was left sided and posterior, being simply a trabecular pouch with neither inlet nor outlet portions, both great arteries arising from the right ventricle. In these four the aorta was posterior and to the right in two, and anterior and to the right in the other two. This was in contrast to the three patients with atrioventricular concordance and imperforate left valve membranes. In these three

Table 1 *Catheter data*

Case No.	*Age*	*PAP*	*AOP*	*QP*	*LAP*	*RAP*	*% O_2 SAT*	*PVR*
1	4 wk	77	—	3·5	25	4	70	8·8
2	2 y	96	90	—	—	3	70	—
3	1 y	45	100	5·9	11	11	74	0·8
	8 y	28	90	1·7	20	7	88	4·1
4*	19 mth	80	80	7·0	11	3	75	6·1
5	16 y	—	125	—	5	3	76	—
6	6 wk	—	100	—	19	19	—	—
	3 y	—	80	3·6	5	5	78	—
	6 y	—	100	—	19	19	79	—
	10 y	—	90	4·6	9	9	86	—
7*	8 mth	85	83	2·4	9	5	84	8·4
8*	2 mth	84	76	2·4	—	11	65	—
9†	6 wk	59	70	5·6	9	4	74	5·3
	5 mth	80	62	3·4	17	12	68	12·0
10	3 wk	40	65	2·1‡	11	4	88	6·5
11	6 wk	76	76	2·6	27	3	64	8·0
12	3 wk	—	75	5·2	21	9	70	—
13	4 wk	—	—	1·1	25	3	—	—
14	5 y	85	90	9·1	18§	5	91	5·7
15	5 wk	80	85	5·2	—	9	94	6·2
	7 wk	35	60	5·4	20	40	81	1·3
	6 y	25	100	3·6	11	11	80	—
	10 y	47	75	2·5	7	5	77	—
16	6 mth	66	75	—	16	12	84	—
	3 y	68	87	3·2	—	2	76	12·0
17	3 mth	55	—	5·1	4	4	—	5·4

* Postoperative death. † Operative death. § Pulmonary artery wedge. ‡ Effective pulmonary flow.
PAP, pulmonary artery pressure; AOP, aortic pressure; QP, pulmonary flow; LAP, left atrial pressure; RAP, right atrial pressure; PVR, pulmonary vascular resistance.

patients the hypoplastic left ventricle gave rise to the aorta, the ventricles and great arteries being normally related.

In the three remaining patients with absent left atrioventricular connection, the right atrium was connected with a single ventricular chamber of indeterminate type, both great arteries arising from this ventricle. The aorta was directly anterior in one patients, in the left posterior position in one patient, and in the right posterior position in the remaining case.

Associated anomalies in the group as a whole included persistent ductus arteriosus in one case, juxtaductal coarctation in two cases, partial anomalous venous drainage with the right pulmonary veins entering the superior vena cava in two cases, left superior vena cava in four cases, and stenosis of the superior vena cava in one case.

Balloon atrial septostomy was performed in four patients at the end of cardiac catheterisation.

SURGERY

The operations in the sequence in which they were performed are outlined in Table 2.

Atrial septectomy was performed in nine patients. In those patients presenting in the first three months of life, atrial septectomy was performed by inflow

Table 2 *Surgical precedures performed in each patient*

Case No.	*Age*	*Procedure(s)*
1	4 wk	Septectomy, PA banding
2	2 y	Septectomy, PA banding, relief of SVC obstruction
3	8 y 7 mth	Septectomy, pulmonary valvotomy
4*	5 y	Septectomy
5	16 y	Modified Fontan's
6	5 y	Waterston
	5 y 4 mth	Potts
7*	9 mth	PA banding
8	3 mth	Septectomy
9	4 wk	Resection coarctation division, PDA ligation, PA banding
	2 mth	Septectomy
10†	2 mth	Resection coarctation
	5 mth	PA banding
	5½ mth	Septectomy
11	3 wk	Septostomy
	4 wk	PA banding
	5 wk	Septectomy
12†	6 wk	Septostomy
	6 wk	Septectomy
	8 wk	PA banding
13	3 wk	Septostomy
	6 wk	PA banding
14	4 wk	Septostomy
15	—	—
16	6 mth	PA banding
17	4 mth	PA banding

* Operative death. † Postoperative death.

occlusion. In older patients septectomy was performed using cardiopulmonary bypass. This is our preferred method in these patients because a complete septectomy can be performed and it allows detailed inspection of the intracardiac anatomy. Banding of the pulmonary artery was performed in ten patients, resection of a juxtaductal coarctation was performed in two patients, and ligation of a persistent ductus arteriosus in one. One patient had a complete stenosis of the superior vena cava and partial anomalous venous drainage, and a Goretex graft was inserted between the superior vena cava and the right atrium at the time of atrial septectomy.

A more physiologically corrective procedure was performed in one girl. She presented at the age of 16 years with increasing cyanosis, dyspnoea on exertion, and a past history of two cerebrovascular accidents. Cardiac catheterisation and angiography disclosed absent left atrioventricular connection in association with a univentricular heart of left atrioventricular type with concordant ventriculoarterial connections and normally related great arteries (Holmes heart with mitral atresia).[8] At operation she was found to have a large secundum atrial septal defect. An interatrial baffle was constructed to divert the pulmonary venous blood through the right atrioventricular valve. An aortic homograft valve was then placed at the junction of the inferior vena cava and the right atrium. The rudimentary right ventricular chamber was opened, the outlet foramen closed, and a 1·8 cm preclotted woven Dacron tube placed between the right atrial appendage and the outlet chamber (Fig. 2).

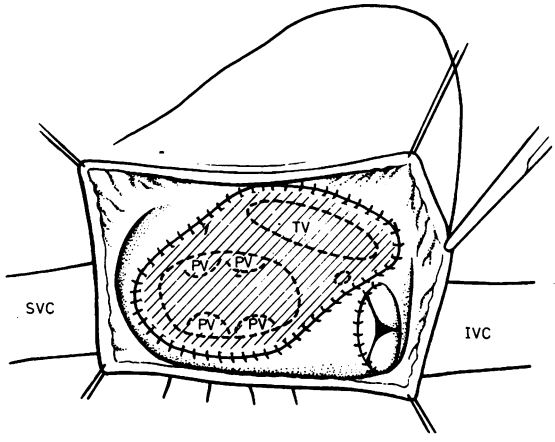

Fig. 2 *An intra-atrial baffle has been placed to direct blood from the four pulmonary veins (PV) into the orifice of the right atrioventricular valve (TV). Systemic venous return from the superior vena cava (SVC) and inferior vena cava (IVC) is directed into the pulmonary artery by a modified Fontan's procedure, not shown in the drawing.*

HOSPITAL MORTALITY

There were five hospital deaths. Three followed septectomy and two followed pulmonary artery banding. All three patients who died after atrial septectomy had evidence of raised pulmonary vascular resistance and in addition one also had evidence of subaortic obstruction. Of the two patients who died after pulmonary artery banding, one had atrioventricular valve regurgitation and the other had a cardiac arrest after accidental removal of the endotracheal tube and suffered irreversible brain damage.

FOLLOW UP

Systemic oxygen saturation was increased by septectomy in all but one patient, including those patients who had a simultaneous pulmonary artery banding (Table 3). One patient had a particularly dramatic increase in oxygen saturation from 64 to 95%. Pulmonary artery banding was subsequently required and in retrospect should have been performed at the time of septectomy. Of the six patients who have had a septectomy, five have also had pulmonary artery banding. Follow up in these patients ranges from five to 150 months with a mean of four years. None of the patients had more than minimal reduction in exercise tolerance: two are pink at rest and the other four are minimally desaturated.

One patient underwent balloon septostomy seven years ago. He is at present well but has clinical evidence of pulmonary hypertension. In retrospect he should have had pulmonary artery banding.

Another patient who underwent balloon septostomy followed by pulmonary artery banding is well but has electrocardiographic evidence of left atrial hypertrophy.

Another patient catheterised 10 years ago at the age of 5 years had neither balloon septostomy nor pulmonary artery banding in spite of the fact that her pulmonary artery pressure was at systemic level, and there was a gradient of 13 mmHg between the right

Table 3 *Absent left atrioventricular connection*

% O_2 Saturation	
Before septectomy	*After septectomy*
81	77*
70	88†
80	91*
64	95‡
70	90†
88	91
75	87§
76±10%	89±6%

* Previous banding. † PA banding.
‡ Banding required 2 weeks later. § Postoperative death.

atrial and pulmonary artery wedge pressure. She now has clinical evidence of pulmonary hypertension.

A further patient who had pulmonary artery banding at the age of 6 months, did not have atrial septectomy because the interatrial gradient at this time was only 4 mmHg, and now there is clinical evidence of left atrial hypertension; when recently recatheterised the left atrium could not be entered.

The patient who underwent a modified Fontan's procedure at the age of 16 required a further operation to relieve stenosis at the distal end of the conduit produced by pseudoendothelial encroachment. She is now aged 20 and leads an active, virtually normal, life.

There have been no late deaths in this series.

Discussion

Left atrioventricular valve atresia in patients with situs solitus is most frequently associated with atresia of the aortic root.[1–3] In this situation systemic and coronary blood flow depends upon a persistent ductus arteriosus and most patients die in the first few days of life. In contrast, when atresia of the left atrioventricular valve is associated with a patent aortic root, survival beyond the neonatal period is much more frequent. If suitable palliative procedures enhance survival in this group, then more corrective surgical procedures can be envisaged during childhood.

Mickell *et al.*[6] recently reviewed their clinical experience with this group of patients, and described how palliative procedures should be designed first to optimise pulmonary blood flow and second to relieve left atrial hypertension. Our findings confirm their observation that left atrial hypertension caused by inadequate interatrial communication can be identified on serial cardiac catheterisations in the majority of patients. In both series, such hypertension was progressive but was not always associated with obvious clinical deterioration. Relief of left atrial hypertension, by balloon atrial septostomy in infancy or atrial septectomy in older patients, should therefore be provided as soon as a restrictive interatrial communication is identified. Banding of the pulmonary artery should be performed in those patients without naturally occurring pulmonary outflow tract obstruction, to prevent the development of pulmonary vascular disease. There is a significant early mortality after palliative procedures. This may well be related to the fact that pulmonary vascular disease had already developed in some cases, thus emphasising the necessity for early surgical intervention.

The need for continuous reassessment and recatheterisation, pointed out by Mickell *et al.*,[6] is well illustrated by three patients in our series. In two, non-restrictive interatrial communication became restrictive in later life while the other developed a restrictive interatrial communication after balloon septostomy.

There was too few patients in our series with pulmonary outflow tract obstruction to support the conclusion of Mickell *et al.*[6] that the prognosis was best in those with mild to moderate pulmonary stenosis. Having performed palliative surgery to provide to the best of our ability the ideal situations outlined above, all the patients in this series bar two have minimal limitation of exercise tolerance and are leading virtually normal lives. The two outstanding patients are severely restricted. One has severe pulmonary outflow tract obstruction and one has developed a restrictive interatrial communication and is awaiting surgery.

The possibilities for more "corrective" surgery are illustrated by our case managed successfully by a modified Fontan's procedure, a procedure not to the best of our knowledge previously performed for this anomaly but suggested recently by Rao *et al.*[7] Our patient is leading a normal and active life at the age of 20, four years after operation. It must be said, however, that the particular morphology in this case, though ideal for the operation, is a rare combination. There was ventriculoarterial concordance in association with a univentricular heart of left ventricular type. This permitted the rudimentary right ventricular chamber to be incorporated into the pulmonary circuit as is possible in classical tricuspid atresia.[9] More usually, as illustrated in our series and other reported cases,[10] the ventriculoarterial connection is discordant when absent left atrioventricular connection is found in association with a univentricular heart of left ventricular type. If "corrective" surgery were envisaged in these cases, it would be necessary, after rerouting of atrial blood, to connect the newly constructed right atrium directly to the pulmonary arteries. Similar procedures will probably be required when absent left atrioventricular connection is found in association with a univentricular heart of right ventricular type. In the four hearts of this type in our series both great arteries arose from the right ventricular main chamber, the rudimentary left ventricular chamber being simply a trabecular pouch of no value for incorporation within either circuit during reconstructive surgery. Similarly, in those patients with absent left connection in association with sole ventricular chambers of indeterminate type, "corrective" surgery would entail septectomy, division of the common atrial chamber thus created, and connection of a newly created right atrium directly to the pulmonary arteries. The success of such operations may therefore be limited, since

propulsion of the pulmonary circulation will depend on a newly created systemic venous atrium with less than its normal musculature. Only experience will show whether such a modified Fontan procedure is possible. What can be stated is that for such an operation to have any chance of success, the pulmonary vascular resistance must be low.[11] This re-emphasises importance of early palliative procedures to relieve left atrial hypertension and to curtail excessive pulmonary blood flow.

It is also highly likely that a modified Fontan procedure would be the most suitable "corrective" procedure in those cases which have imperforate left atrioventricular valves in association with atrioventricular concordance. In such hearts in our series, though the left atrium was connected to the hypoplastic left ventricle, the valve membranes and their tension apparatus were grossly hypoplasitc. It did not seem feasible to insert a prosthetic valve in this small left atrioventricular orifice even if it proved possible to remove the imperforate valve membrane. Equally, the left ventricle itself seemed so hypoplastic as to be incapable of supporting the circulation if septation had been attempted in combination with re-establishment of left atrial-left ventricular continuity. Thus, as with the univentricular hearts, the best chance of success, if any success is possible, seems to lie in atrial septectomy, construction of a new atrial septum, and use of the modified Fontan procedure. The aorta would then receive pulmonary venous blood via the right ventricle and the ventricular septal defect. As with the majority of hearts in our series, the possibility of success of such a procedure is highly speculative.

The above emphasises that in most instances our practice of dividing left atrioventricular valve atresias into the morphological categories of absent connection and imperforate valve confers little practical clinical value. The entity is coloured most strongly by the lack of egress from the left atrium whatever its anatomical substrate. None the less, recognition of the different morphological possibilities is of significance in determining the possible surgical options as in our one case where a modified Fontan procedure was feasible. Equally, though we have yet to see such a case, hearts may be encountered in which an imperforate membrane is of sufficient size to justify its surgical removal and replacement with a prosthetic valve. Then it would be essential to distinguish the presence of the membrane.

At present, it can be said that cases of left atrioventricular valve atresia in the presence of a patent subaortic outflow tract can be palliated with an acceptable degree of success. Whether more "corrective" procedures will become available for these palliated patients remains to be established, but at least in our experience the palliation in itself has provided an acceptable quality of life.

References

1 Redo SF, Farber S, Gross RE. Atresia of the mitral valve. *Arch Surg* 1961; **82**: 696–712.
2 Eliot RS, Shone JD, Kanjuh VI, Ruttenberg HD, Carey LS, Edwards JE. Mitral atresia. A study of 32 cases. *Am Heart J* 1965; **70** 6–22.
3 Teller WM. Congenital mitral atresia. *Am Heart J* 1958; **56**: 304–12.
4 Watson DG, Rowe RD, Conen PE, Duckworth JWA. Mitral atresia with normal aortic valve. Report of 11 cases and review of the literature. *Pediatrics* 1960; **25**: 450–67.
5 Moreno F, Quero M, Diaz LP. Mitral atresia with normal aortic valve. A study of 18 cases and a review of the literature. *Circulation* 1976; **53**: 1004–10.
6 Mickell JJ, Mathews RA, Park SC, *et al.* Left atrioventricular valve atresia: clinical management. *Circulation* 1980; **61**: 123–7.
7 Rao PS, Kulangara RJ, Moore HV, Strong WB. Syndrome of single ventricle without pulmonary stenosis but with left atrioventricular valve atresia and interatrial obstruction. *J Thorac Cardiovasc Surg* 1981; **81**: 127–30.
8 Quero M. Atresia of the left atrioventricular orifice associated with a Holmes heart. *Circulation* 1970; **42**: 739–44.
9 Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971; **26**: 240–8.
10 Tandon R, Edwards JE. Tricuspid atresia. A re-evaluation and classification. *J Thorac Cardiovasc Surg* 1974; **67**: 530–42.
11 Choussat A, Fontan F, Besse P, Vallot F, Chauve A, Bricaud H. Selection criteria for Fontan's procedure. In: Anderson RH, Shinebourne EA, eds. *Paediatric cardiology*. London: Churchill Livingstone, 1977: 566-99.

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